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Case report

Langerhans cell histiocytosis presenting with complicated pneumonia, a case report

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ABSTRACT

We describe a 2 ½ year old boy presenting with fever, abdominal pain and splinter haemorrhages of the nails. On further examination there were signs of pneumonia with pleural effusion. This was treated with mini-thoracotomy, drainage and intravenous antibiotics. Further diagnostic workup for underlying causes showed diffuse cystic lung disease, suggestive of Langerhans cell histiocytosis. This was confirmed on pathology specimens, which showed Langerhans cells in lung tissue, nail bed and skin biopsy samples, indicating multisystem Langerhans cell histiocytosis. The patient was treated with Prednisone and Vinblastin according to the LCH-III guidelines. In this case report we give a brief description on cystic lung disease in children, Langerhans cell histiocytosis and associated nail abnormalities.

1. Introduction

Langerhans cell histiocytosis (LCH) is a rare disease characterized by monoclonal proliferation of dendritic-cell related histiocytes (Langerhans cells). These histiocytes have destructive behaviour for the surrounding tissue which they infiltrate. Among the organs often involved are the skeletal system, skin, thyroid gland and risk organs like liver, lung, spleen and the haematopoietic system.

In this case we present a young toddler primarily presenting with signs of a complicated pneumonia, as the presenting sign of underlying systemic disease. Although lung involvement is frequently seen in multisystem LCH, it is not often the presenting sign.

2. Patient presentation

A 2 ½ -year-old boy was referred to the paediatric outpatient department for acute fever, abdominal pain, and decreased oral intake. Furthermore, he had linear splinter haemorrhages of the nails for some weeks. Previous medical history was unremarkable besides a recent tonsillectomy. Physical examination showed an ill child with fever, tachypnoea (breathing rate 36/min), tachycardia (heart rate 165/min) and oxygen saturation in room air between 89 and 93%. With oxygen supplementation via nasal cannula of 1 L/min, oxygen saturation was 97–98%. Blood pressure was 121/76 mmHg. On auscultation there was decreased air-entry over the left hemithorax, pleural friction rub, and hepatomegaly. Laboratory data revealed elevated C-reactive protein of 333 mg/l (reference range <10 mg/l), erythrocyte sedimentation rate was 37 mm/h (reference range 3–13 mm/h), platelet count 573 $\times$ 10^9/l (reference range 150–450 $\times$ 10^9/l), white blood cell count 24.6 $\times$ 10^9/l (reference range 4.0–10.0 $\times$ 10^9/l). Differential blood count: neutrophils 86.6%, immature neutrophils 6%, lymphocytes 7.6%, monocytes 5.3%, eosinophils 0.3%, basophils 0.2%.

The chest radiograph revealed a basal consolidation in the left lower lobe and opacification along the lateral chest wall. Moreover, there was some right-sided displacement of the heart and mediastinum. This was suspect of pulmonary consolidation with pleural effusion. Additionally, the right lung and left upper lobe demonstrated an evident reticulonodular pattern (Fig. 1).

The diagnosis of pneumonia with pleural effusion was made. This was confirmed with ultrasound. At this point the decision was

Abbreviations: LCH, Langerhans cell histiocytosis; HRCT, High resolution computed tomography.

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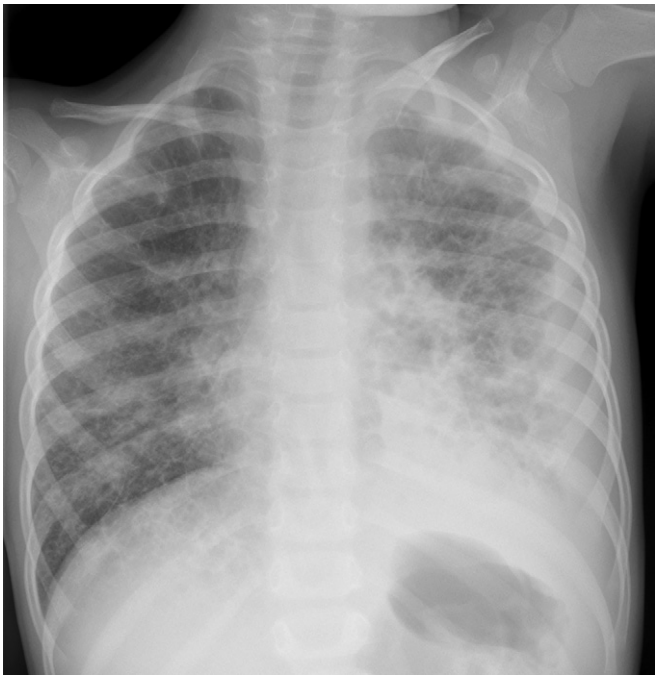

Fig. 1. Chest radiograph at presentation. Basal consolidation in the left lower lobe, opacification along the left lateral wall and mediastinal shift, indicating pneumonia and pleural effusion. Reticulonodular pattern in right lung and left upper lobe.

made not to perform a diagnostic pleural tap, but start treatment with broad-spectrum antibiotics, amoxicillin-clavulanic acid and gentamicin. Blood cultures remained negative. After initial improvement, the patient deteriorated after four days with dyspnoea and increased oxygen need. Chest ultrasound showed increased pleural effusion and progressive organisation of the effusion. Bacterial endocarditis was ruled out with negative blood cultures and a normal cardiac ultrasound. A mini-thoracotomy was performed with decortication of the left lung and placement of a pleural drain. Pleural fluid chemistry showed signs of pleural exudate (pH 6.92, glucose <0.6 mmol/l, protein 35.7 g/L, lactate dehydrogenase 2677 U/l)[1] Antibiotics were switched to flucloxacillin, gentamicin and clindamycin. Bacterial cultures of pleural fluid and blood remained negative. Afterwards, analysis of pleural fluid with polymerase chain reaction (16S-PCR) determined *Streptococcus pneumoniae* as the causative pathogen. The patient fully recovered within 10 days, had no oxygen need and was dismissed from hospital care with oral antibiotics.

Since the chest radiograph showed not only pneumonia and pleural effusion, but also interstitial abnormalities, a thorough diagnostic workup was performed to rule out underlying causes of pulmonary disease. The inflammatory markers had normalized. Serological tests for viral, atypical and bacterial pathogens were negative. Sweat test was negative. Tuberculin skin test was negative.

The immunological survey was normal. The patient had been vaccinated with a heptavalent pneumococcal conjugate vaccine (PCV-7, Prevenar®). There were normal pneumococcal antibody levels. Subtyping of the pneumococcal strand was not possible, since it was detected with 16S-PCR, not by culture.

Further imaging was planned to be performed after full recovery of the pleural empyema. Eight weeks after full recovery the chest radiograph was still abnormal with a reticulonodular pattern and features of honeycombing (Fig. 2). Therefore, a high resolution Computed Tomography (HRCT) was performed. The HRCT of the thorax revealed numerous bilateral cysts of different size and varying wall thickness (Fig. 3). There were no signs of emphysema or bronchiectasis. Hence, Langerhans cell histiocytosis (LCH) was suspected.

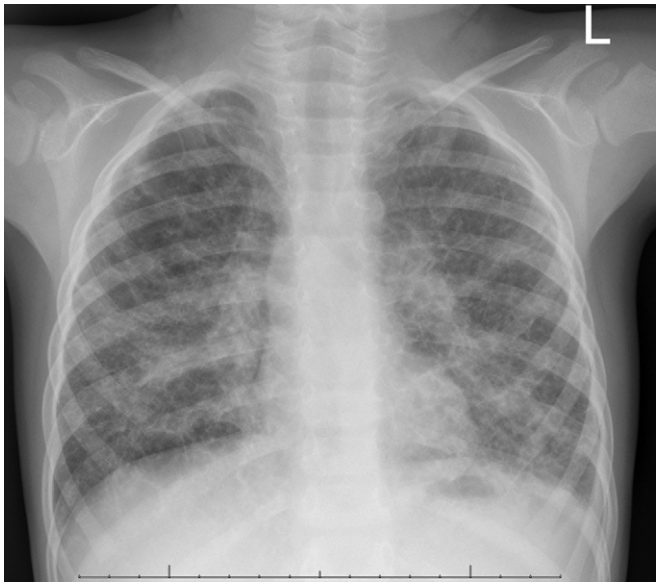

Fig. 2. Chest radiograph, performed eight weeks after recovery, revealing persistent reticulonodular pattern with features of honeycombing.

Further diagnostic testing was performed to confirm the diagnosis. Abdominal ultrasound was normal, with no signs of hepatic involvement or hepatomegaly and normal liver function tests. The initial presentation with hepatomegaly had been caused by diagfragmatic flattening due to pulmonary hyperinflation.

Full body X-ray series showed no osteolytic lesions indicative for LCH. Under general anaesthesia an open lung biopsy, skin biopsy, nail bed biopsy, bone biopsy and bone marrow aspiration was performed.

The lung biopsy showed foci of histiocytes, mixed with eosinophilic granulocytes, with a stellar distribution. In these foci cysts were developing. CD1a staining was highly positive and S100 staining mildly positive, indicative of Langerhans Cell Histiocytosis. Skin- and nail bed specimens also showed presence of CD1a-positive cells. Bone marrow aspiration was negative.

The patient was treated following the LCH-III protocol (Histiocyte Society) with Vinblastin and Prednison. After six weeks, the patient responded well to chemotherapy at the first course evaluation. Nail and skin abnormalities improved, and chest CT remained stable. Bronchoalveolair lavage at 12 weeks treatment showed less

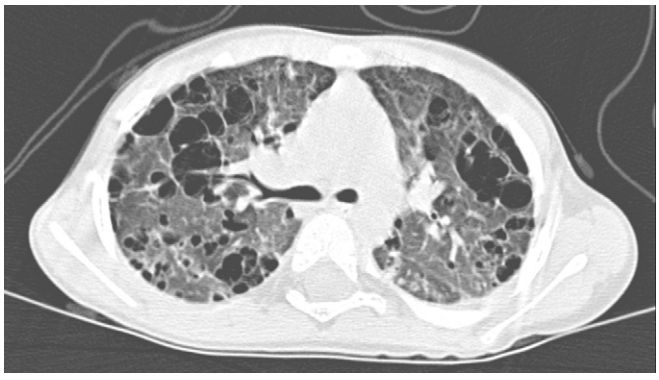

Fig. 3. High resolution CT. Diffuse bilateral cysts of varying size and wall thickness.