Pulmonary Langerhans cell histiocytosis (PLCH): a new UK register

Abstract
Pulmonary Langerhans cell histiocytosis (PLCH) is a rare interstitial lung disease of unknown aetiology. We aimed to characterise a UK-wide cohort of patients with PLCH and compare diagnostic and management methods in specialist and non-specialist centres. 106 cases (53 hospitals) identified. Complete data received in 67 cases (53.7% female, age 37.1±14.4 years). 96% current or ex-smokers. Treatment; smoking cessation (79%), corticosteroids (30.6%), cytotoxic therapy (26.9%) and lung transplant (6%). Patients at specialist centres received cytotoxic drugs more often (p=0.0001) and survival appeared higher. This dataset indicates a more even gender distribution than previously documented. It suggests variation in clinical management and outcomes achieved dependent on clinical experience.

One hundred and six patients (17 deceased, 8 lost to follow-up) were initially identified from the British Thoracic Society Orphan Lung Disease (BOLD) database and subsequently through advertisements in the eBritish Thoracic Society (BTS) bulletin, at BTS meetings, the BTS BOLD conference and by contacting all UK interstitial lung disease leads. Demographic and clinical data were collected with consent (Ethics no: 08/H0104/98) by post, from individual patients, their respiratory clinicians and their general practitioners.

One hundred and six patients (17 deceased, 8 lost to follow-up) were initially identified from 53 centres. Of these, sufficient data to allow analysis was available for 67 cases (mean age at presentation 37.1±14.4 years, 53.7% female).

The main presenting symptoms were dyspnoea (78%) and cough (63%); 80.7% had a normal respiratory examination. Mean pack year smoking history was 19.9 (SD) FEV₁ 72.3±25.3%, FVC 86.2 ±23.3%. Of the patients 38% had an obstructive pattern and 16% restrictive. Of the cases, 70.5% had transfer factor (TLC/VT) <70% predicted.

Diagnosis was made on surgical biopsy in 61% of cases. Treatment received included; smoking cessation (79%), corticosteroids (30.6%), cytotoxic therapy (26.9%), pleural intervention (16.4%) and lung transplant (6%).

Twenty-seven of our 67 patients were managed in a specialist centre. Patients were significantly more likely to have had an echocardiogram (p=0.01) and receive cytotoxic drugs if managed in a specialist centre (p=0.0001). No difference was detected in the number of open lung biopsies performed (p=0.8). Survival appeared higher in those treated at a specialist centre (p=0.03) (see table 1).

In conclusion, this population-wide analysis of patients with PLCH in the UK has shown similarities with previously described cohorts from other countries. It has reconfirmed a potential change in disease distribution with female preponderance and its apparent association with cigarette smoking. It has indicated that despite advances in CT, a high proportion of patients still require an open lung biopsy for diagnostic confirmation. Finally, it has also identified the variation in the treatment options available for patients depending on where their disease is managed and suggests the need to produce guidance for all clinicians to remove potential treatment inequalities.

Our register is still open to recruitment and we are keen to hear from any clinicians with patients with PLCH.

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Table 1 Comparison of demographics, mode of diagnosis and clinical management between patients managed in a specialist centre versus a non-specialist centre

<table>
<thead>
<tr>
<th>Outcome</th>
<th>N (%)</th>
<th>Non-specialist</th>
<th>Specialist</th>
<th>χ² p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Deceased</td>
<td>9 (22.5)</td>
<td>1 (3.7)</td>
<td>8 (29.6)</td>
<td>0.03*</td>
</tr>
<tr>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Smoking cessation</td>
<td>25 (62.5)</td>
<td>21 (77.8)</td>
<td>0.19</td>
<td></td>
</tr>
<tr>
<td>Steroids</td>
<td>10 (25)</td>
<td>9 (33.3)</td>
<td>0.46</td>
<td></td>
</tr>
<tr>
<td>Cytotoxic drugs</td>
<td>4 (10)</td>
<td>14 (51.8)</td>
<td>0.0001*</td>
<td></td>
</tr>
<tr>
<td>Lung transplantation</td>
<td>1 (2.5)</td>
<td>0 (0)</td>
<td>0.84</td>
<td></td>
</tr>
<tr>
<td>Diagnosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bronchoscopy</td>
<td>9 (22.5)</td>
<td>8 (29.6)</td>
<td>0.5</td>
<td></td>
</tr>
<tr>
<td>HRCT scan</td>
<td>28 (70.0)</td>
<td>27 (100)</td>
<td>0.05*</td>
<td></td>
</tr>
<tr>
<td>Lung biopsy</td>
<td>19 (47.5)</td>
<td>12 (44.4)</td>
<td>0.8</td>
<td></td>
</tr>
<tr>
<td>Investigation</td>
<td>10 (25)</td>
<td>15 (55.6)</td>
<td>0.01*</td>
<td></td>
</tr>
</tbody>
</table>

* Significant result (p<0.05).

HRCT, high resolution computed tomography.
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Contributors Study concept and design: JS and RHM; Acquisition of data, analysis and interpretation of data: JS, RHM and HIA; Statistical analysis: RHM and JS; Drafting of the manuscript and critical revision of the manuscript for intellectual content: JS, RHM, NMF, HMB, HIA, MH and TMM; Study supervision: JS.

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