The management of adrenal cell carcinoma in a single tertiary centre: A 25 year experience

Nicole Goff, Claire Hughes, Harshini Katugampola, Imran Mushtaq, Peter Hindmarsh, Catherine Peters, Caroline Brain, Mette Jorgensen, Mehul Dattani

Great Ormond Street Hospital, London, United Kingdom, Royal London Hospital Barts Trust, London, United Kingdom, Queen Mary, University of London, London, United Kingdom, UCL Great Ormond Street Institute of Child Health, London, United Kingdom, University College London Hospital, London, United Kingdom

Background

Adrenal cortical carcinoma (ACC) in children is rare and aggressive, with the mainstay of treatment being surgical resection. Although, there have been recent improvements in outcomes with chemotherapy, the prognosis is still quite poor and often patients present with advanced disease. Focus on endocrine management has not been previously described.

Objective

- Further characterise
- Presenting features and biochemical markers to support earlier diagnosis
- Strategies to manage refractory hypertension
- Describe longer term endocrine management in this cohort

Methods

Retrospective review of patient records at a single centre between 1996 and 2021 of patients with an ACC.

Results

<table>
<thead>
<tr>
<th>Presentation</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Virilisation</td>
<td>29</td>
</tr>
<tr>
<td>Pubic hair</td>
<td>25</td>
</tr>
<tr>
<td>Hypertension</td>
<td>20</td>
</tr>
<tr>
<td>Acne</td>
<td>17</td>
</tr>
<tr>
<td>Weight gain</td>
<td>17</td>
</tr>
<tr>
<td>Cushingoid</td>
<td>15</td>
</tr>
<tr>
<td>Clitoromegaly</td>
<td>9</td>
</tr>
<tr>
<td>Hypertrichosis</td>
<td>8</td>
</tr>
<tr>
<td>Precocious Puberty</td>
<td>7</td>
</tr>
<tr>
<td>Palpable Mass</td>
<td>7</td>
</tr>
<tr>
<td>Greasy hair</td>
<td>7</td>
</tr>
<tr>
<td>Hoarse voice</td>
<td>4</td>
</tr>
<tr>
<td>Abdominal Distension</td>
<td>4</td>
</tr>
<tr>
<td>Seizures</td>
<td>1</td>
</tr>
<tr>
<td>Gynaecomastia</td>
<td>1</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Management</th>
<th>Detail</th>
</tr>
</thead>
<tbody>
<tr>
<td>Refractory Hypertension</td>
<td>Metrapone (n=3) Ketaconazole (n=3)</td>
</tr>
<tr>
<td>Surgical Resection</td>
<td>Evolved ovtetime to become laparoscopic</td>
</tr>
<tr>
<td>Adjuvant Chemotherapy</td>
<td>32% of patients received Mitotane (n=8)</td>
</tr>
<tr>
<td>Hydrocortisone</td>
<td>Most treated with IV infusion All were discharged on hydrocortisone replacement (10mg/m2/day; duration 0.1-8y, median 1y) Those treated with mitotane required higher doses (15-17mg/m2/day)</td>
</tr>
<tr>
<td>Fludrocortisone</td>
<td>3 patients (all associated with mitotane)</td>
</tr>
<tr>
<td>Thyroxine</td>
<td>2 patients (all associated with mitotane)</td>
</tr>
</tbody>
</table>

Key messages

- The most frequent presentations are virilisation, hypertension, cushingoid appearance, and rarely gynaecomastia
- This highlights the essential role of genetic diagnosis in influencing disease progression and treatment
- Hydrocortisone
  - Our practice has evolved to include post-operative intravenous hydrocortisone infusion in all patients to mitigate the effect of the rapid fall in cortisol
  - The duration of requirement in this cohort was highly variable
- Future Research Direction
  - Newer agents such as pasireotide or mifepristone to decrease cortisol concentrations in those with refractory hypertension, as they may have fewer side-effects and reduce morbidity

*Percentage of cases