New onset Sarcoidosis following treatment of Cushing’s Disease

Dr. R Cairns & Dr. L Hall
Department of Diabetes & Endocrinology
Wishaw General Hospital, Scotland, UK
Email: ross.cairns@nhs.net

Introduction

Cushing’s Disease (CD) is caused by pituitary adrenocorticotropic hormone (ACTH) secreting tumours. These tumours are almost always benign in nature and are usually microadenomas by classification with the disease characterised by excessive elevation of glucocorticoid concentrations. [1]

In rare cases the treatment of Cushing’s Disease, with a subsequent reduction in glucocorticoid concentration, may result in the unveling or aggravation of a disease responsive to glucocorticoid therapy. [2][3][4]

We describe a case were a patient was diagnosed with new onset Sarcoidosis following treatment of Cushing’s Disease.

Investigations, management and follow up

A diagnosis of Cushing’s Disease was made and the patient underwent an uncomplicated left sided transphenoidal hemi-hypophsectomy. The results of the initial biochemistry, radiological and subsequent pathology investigations can be viewed in table 1. On postoperative day three the patient’s serum cortisol was < 30 nmol/l and therefore replacement hydrocortisone was commenced. Despite an initial tranquil postoperative convalescence the patient was frequently admitted and readmitted to hospital with a systemic illness of unclear aetiology.

Over these hospital admissions the patient complained of progressive myalgia, arthralgia, lethargy and shortness of breath but with no clear evidence of sepsis or overt hypocortisolis. However, the patient was felt to improve clinically with intravenous glucocorticoids and empirical antibiotic therapy. During one of the hospital admissions a CT Thorax was repeated and this showed new ipsilateral parahilar and hilar lymphadenopathy. The patient’s serum ACE level was found to be elevated, 107 IU/L and Sarcoidosis granulomas in keeping with a diagnosis were demonstrated on eventual hilar biopsy. The patients clinical condition has since spontaneously improved: her respiratory function tests and chest radiograph showed no evidence of disease progression and the serum ACE had fallen without the need for high dose corticosteroids.

Table 1: Investigations for CD

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Result 1</th>
<th>Result 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>24-hour Urine Free Cortisol (UFC)</td>
<td>540 nmol/d</td>
<td>624 nmol/d</td>
</tr>
<tr>
<td>Overnight Dexamethasone Suppression Test (ODST)</td>
<td>671 nmol/L</td>
<td>791 nmol/L</td>
</tr>
<tr>
<td>Low Dose Dexamethasone Suppression Test (LDST)</td>
<td>105 nmol/L</td>
<td>N/A</td>
</tr>
<tr>
<td>High Dose Dexamethasone Suppression Test (HDST)</td>
<td>63 nmol/L</td>
<td>N/A</td>
</tr>
<tr>
<td>Morning ACTH concentration</td>
<td>7 IU/L</td>
<td>8 IU/L</td>
</tr>
<tr>
<td>Bilateral Inferior Petrosal Sinus Bypassing</td>
<td>Failed</td>
<td>Failed</td>
</tr>
<tr>
<td>CT Thorax, Aldosteron and Pilsic with contrast</td>
<td>No adrenal source for ACTH secretion or other abnormality found</td>
<td></td>
</tr>
<tr>
<td>3T MRI Head</td>
<td>Left sided 3 x 4 x 6 mm interparietal pituitary adenoma</td>
<td></td>
</tr>
<tr>
<td>Pituitary Pathology</td>
<td>Pituitary macroadenoma with positive immunostaining for ACTH</td>
<td></td>
</tr>
</tbody>
</table>

Discussion

- Glucocorticoid responsive illnesses can be unmasked or aggravated following treatment of Cushing’s Disease and it is thought that the rebound immunity that occurs plays a significant role. [5]
- Cases reports have described exacerbation and onset of inflammatory arthropathies, vasculitides, thyroid and granulomatous disease. [2] [4]
- Sarcoidosis has been reported as a rare sequela following treatment of CD but usually presents with skin manifestations such as erythema nodosum. [2]
- This case is unusual in that dermatological signs were absent and that only a generalised systemic illness with hilar and mediastinal lymphadenopathy was seen.

Conclusion

- The unveling of a glucocorticoid responsive disease post treatment for Cushing’s Disease is rare.
- The convalescent period following treatment for Cushing’s Disease can be stormy and consideration must occasionally be given to unusual diagnoses.

References