with conventional radiotherapy. Only four of our 19 patients, however, were younger than 20 years of age when irradiated.

The development of Nelson’s syndrome is the most serious complication of bilateral adrenalectomy, and two recent series reported incidences of 29% and 28%. Though conventional radiotherapy does not prevent the development of Nelson’s syndrome, it seems to reduce its incidence. Whether low dose radiation treatment will exert a similar beneficial effect in patients who subsequently require adrenalectomy is not known. Five of our patients had an adrenalectomy and were followed up for two to eight years with no evidence of Nelson’s syndrome. In conclusion, we recommend that patients with Cushing’s disease be offered initial treatment with low dose radiation treatment, especially in centres where neurosurgical skill in microadenomectomy is not readily available. Those patients who do not have remission of their disease by one year after radiotherapy should be considered for bilateral adrenalectomy. We believe that by this approach at least one third of patients with Cushing’s disease have a chance of cure with a relatively simple and non-invasive treatment, which in our experience does not have any complications.

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References


Lymphadenopathy and selective IgA deficiency

M A H FRENCH

Abstract

Four men presented with unexplained lymphadenopathy. Three had a history of recurrent respiratory infections for several years, and two had lymph node or hepatic granulomas. None was noted to have symptoms of immunodeficiency at the time of presentation.

In one patient routine direct immunofluorescence study failed to detect IgA, and immunological investigations were therefore conducted in the rest. In all patients the findings were similar and characterised by a severe deficiency of IgA.

In the absence of a more serious cause selective IgA deficiency may be enough to explain “idiopathic” lymphadenopathy.

Introduction

Lymphadenopathy is a common clinical problem with a multitude of causes including, on rare occasions, immunodeficiency syndromes. A list of cases has been suggested by virtue of common variable hypogammaglobulinaemia in a minority of patients; lymphadenopathy has not been associated with selective IgA deficiency. The four cases reported here suggest that it may not only be associated with IgA deficiency but may be a presenting feature.

Case histories

CASE 1

A 29 year old man had been investigated on three occasions over five years because of recurrent axillary lymphadenopathy and nocturnal sweats. On each occasion a skin node biopsy had shown follicular hyperplasia and granulomas without evidence of mycobacterial or fungal infection. Thrombocytopenia had also been present, which

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Results of immunological studies (normal values given in parentheses)

<table>
<thead>
<tr>
<th>Serum immunoglobulins (g/l)</th>
<th>IgG subclasses (g/l)</th>
<th>IgG tetanus toxoid antibody (mg/l)</th>
<th>Autoantibodies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case No</td>
<td>IgG (5-16:160)</td>
<td>IgM (0-1-1-5)</td>
<td>IgA (1-3-0)</td>
</tr>
<tr>
<td>1</td>
<td>10:1</td>
<td>1:2</td>
<td>&lt;0-05</td>
</tr>
<tr>
<td>2</td>
<td>8:6</td>
<td>1:9</td>
<td>0-06</td>
</tr>
<tr>
<td>3</td>
<td>11:1</td>
<td>1:3</td>
<td>0-06</td>
</tr>
<tr>
<td>4</td>
<td>16:1</td>
<td>3:2</td>
<td>&lt;0-05</td>
</tr>
</tbody>
</table>

ND = Not done.
* Measured by passive haemagglutination (normal > 1/160).

Methods and results

The table gives the results of the immunological investigations. Serum immunoglobulins were measured by automated immunoprecipitation and, in addition, by radial immunodiffusion using rabbit anticoagulant IgA (Dako, Denmark) for IgA. Measurement of immunoglobulins on several occasions in all patients showed consistent findings. Serum IgG subclasses were measured by radial immunodiffusion using monoclonal antisera and IgG tetanus toxoid antibody by an indirect enzyme linked immunosorbent assay or passive haemagglutination. All sera were examined by conventional techniques for autoantibodies to nuclei, smooth muscle, mitochondria, gastric parietal cells, thyroid microsomes, and thyroglobulin.

Comment

Although lymphadenopathy may be a feature of several immunodeficiency diseases, it is not a recognised complication of selective IgA deficiency. Indeed, many patients with selective IgA deficiency do not have symptoms, although there is an association with autoimmune diseases, allergic diseases, and recurrent respiratory tract infections, which are often minor but sometimes severe. Three of the present four patients had severe respiratory tract infections associated with low IgG tetanus toxoid antibody responses or low serum concentrations of the IgG2 and IgG4 subclasses, or both, suggesting that they were different from most patients with IgA deficiency. Interestingly, two of the patients also had lymph node or hepatic granulomas. Granulomatous disease, usually of the lung but also of the liver, may occur in patients with common variable hypogammaglobulinaemia.

Unlike other immunodeficiency syndromes, in which lymphadenopathy is part of a clinical picture dominated by recurrent infections often associated with other complications, the symptoms of immunodeficiency in these four patients were either minor or overlooked at the time of presentation with lymphadenopathy. In three of the four, however, severe infections were eventually a major feature. Although more serious causes must be excluded, the presence of selective IgA deficiency—particularly with symptomatic immunodeficiency—may be enough to explain "idiopathic" lymphadenopathy.

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References


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