Chronic Diarrhea as a Presenting Manifestation of Common Variable Immunodeficiency

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Received: May 18, 2013; Revised: Jun 20, 2013; Accepted: July 04, 2013

Common variable immunodeficiency (CVID) is one of the primary immunodeficiencies, which usually presents with recurrent bacterial infections, particularly in respiratory and gastrointestinal systems and hypogammaglobulinemia. We present here a case of CVID who was suffering from chronic watery diarrhea since 3 months before admission. Past medical history was uneventful about recurrent infections. No abnormality was revealed on physical examination. After thorough investigations about other possible causes of chronic diarrhea, and based on laboratory data of hypogammaglobulinemia, we considered him as a case of CVID with an isolated manifestation.

Keywords: Common Variable Immunodeficiency; Diarrhea; T cell immunodeficiency

Implication for health policy/practice/research/medical education: This article helps pediatricians for diagnosis of CVID.

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achieved after starting intravenous immunoglobulin administration.

Table 1. Nephelometric Immunoglobulin Analysis Report

<table>
<thead>
<tr>
<th>Immunoglobulin</th>
<th>Patient</th>
<th>Normal Ranges</th>
</tr>
</thead>
<tbody>
<tr>
<td>IgG</td>
<td>0.149</td>
<td>0.5-13 g/L</td>
</tr>
<tr>
<td>IgM</td>
<td>Non-detectable</td>
<td>0.34-2.55 g/L</td>
</tr>
<tr>
<td>IgE</td>
<td>0.1</td>
<td>Up to 68 IU/mL</td>
</tr>
<tr>
<td>IgA</td>
<td>Non-detectable</td>
<td>0.19-2.2 g/L</td>
</tr>
</tbody>
</table>

Table 2. Flow Cytometric Lymphocyte Subset Analyses, Cell Count and Subpopulations

<table>
<thead>
<tr>
<th>Cell Count and Subpopulation</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>White Blood cell Count</td>
<td>8800/mm³</td>
</tr>
<tr>
<td>Neutrophil</td>
<td>40%</td>
</tr>
<tr>
<td>Lymphocyte</td>
<td>53%</td>
</tr>
<tr>
<td>CD3</td>
<td>80.5%</td>
</tr>
<tr>
<td>CD4</td>
<td>10.9%</td>
</tr>
<tr>
<td>CD8</td>
<td>61.3%</td>
</tr>
<tr>
<td>CD56</td>
<td>8.9%</td>
</tr>
<tr>
<td>CD19</td>
<td>9.9%</td>
</tr>
<tr>
<td>CD4/CD8</td>
<td>0.18</td>
</tr>
</tbody>
</table>

3. Discussion

CVID is a diverse group of disorders, which arise from B and T lymphocytes dysfunctions and results in hypogammaglobulinemia (2). CVID has been defined clinically by the presence of recurrent infections and a reduction in IgG (at least 2 SD below the mean), and at least one other immunoglobulin isotypes, as well as by a failure to mount a significant specific antibody response to challenge with vaccination or natural infections (1, 3). CVID usually presents with recurrent sinopulmonary bacterial infections as a result of poor antibody responses and decreased serum immunoglobulins. Infections of the lower respiratory tracts (recurrent pneumonia) often result in bronchiectasis (4). Recurrent respiratory infections affect up to 98% of CVID patients (5). Gastrointestinal manifestations are the presenting symptom of CVID in about 3% of the patients (6). Totally, the gastrointestinal tract is affected in about half of the CVID patients. The major gastrointestinal manifestations of CVID are transient or persistent diarrhea, reported in 21-57% of subjects. Other intestinal presentations include intestinal malabsorption, chronic giardiasis and amebiasis, and atrophic gastritis with pernicious anemia. About 20% of patients with CVID have gut symptoms without an infectious cause (5-9). Although gastrointestinal manifestations are common findings in CVID, they are rare presenting signs and symptoms (as the first presentation) in CVID, especially without concomitant respiratory and gastrointestinal infections. Similarly, in our patient gut infection was never documented on repeated stool examinations and he had never experienced respiratory infections or any other recurrent infections before diagnosis. Actually, chronic non-infectious diarrhea was the single manifestation of CVID in this patient, which is a really rare presentation of CVID. Pater et al. have also recently reported a case of CVID with the rare presentation of chronic diarrhea. They also couldn’t find an infectious origin for his diarrhea, but different from our report, their patient had also previous recurrent infections (10). Another report by Onbasi et al., discusses a 39-year-old patient with CVID, presenting with chronic diarrhea, but giardiasis was found to be the reason of diarrhea. She was also suffering from recurrent infections since childhood (11).

In a study by Aghamohammadi et al. (9), infection, super infection, ulcerative colitis, food-sensitive enteropathy, autoimmune enteropathy, and celiac disease are considered the causes of diarrhea and malabsorption manifestation in these patients.

It is important to consider hypogammaglobulinemia in any patient with a history of chronic diarrhea and patients should undergo a full assessment of immune system including measurement of serum immunoglobulin levels, antibody function evaluation, and B and T-cell subsets enumeration.

The mainstays of treatment for CVID remain replacement immunoglobulin (Ig) and antibiotics for infections and appropriate treatment for the non-infectious complications (7, 9).

Acknowledgements

There is no acknowledgment.

Authors’ Contribution

Amir Hossein Hosseini: Patient’s physician and writing the manuscript; Mehrnaz Mesdaghi: Diagnosis, revising and submitting the manuscript; Ali Akbar Sayyari: Patient’s physician; Zahra Chavoshzadeh: Diagnosis and patient’s physician.

Financial Disclosure

The authors declare that there is no conflict of interest.

Funding/Support

This research/article has not been supported by any institution.

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2. Yong PF, Tarzi M, Chua I, Grimbacher B, Chee R. Common variable


