Stiff person syndrome: a diagnostic and management challenge

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Recommended Citation
Sajjad, Zoya; Abrar, Anam; Ullah, Zafar; Rehman, Aasim; Tariq, Wasim; and Ahmad, Arsalan (2014) "Stiff person syndrome: a diagnostic and management challenge," Pakistan Journal of Neurological Sciences (PJNS): Vol. 9 : Iss. 4 , Article 11. Available at: http://ecommons.aku.edu/pjns/vol9/iss4/11
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This case report is available in Pakistan Journal of Neurological Sciences (PJNS): http://ecommons.aku.edu/pjns/vol9/iss4/11
STIFF PERSON SYNDROME: A DIAGNOSTIC AND MANAGEMENT CHALLENGE

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Date of submission: October 5, 2014, Date of revision: October 22, 2014, Date of acceptance: November 1, 2014

ABSTRACT

Stiff person syndrome (SPS) is a rare neurological disorder characterized by progressive muscle stiffness and rigidity, mostly involving axial muscles, resulting in functional disability. It is associated with elevated anti-Glutamic acid decarboxylase (GAD) antibody levels. Electromyography findings are often diagnostic. We present a case of a 48 years old male, who presented with progressive stiffness and rigidity of axial muscles and limbs. His EMG was consistent with SPS. Anti GAD antibodies were markedly elevated. He was treated with rituximab and has improved significantly. SPS is a difficult diagnosis, usually under diagnosed due to lack of awareness among medical community. There is a dire need to further study the disease and invent better treatment options for patients suffering from SPS.

CASE REPORT

A 48 year old male, known case of Type I Diabetes Mellitus and depression, presented to Neurology Outpatient department with complaints of progressive pain and stiffness of neck, shoulder and abdominal muscles for the past six months. The pain and spasm aggravated with stress and movement and was temporarily relieved on taking Non steroidal anti-inflammatory drugs and muscle relaxants. Initially he did not have any disability and despite stiffness he was able to carry out his daily life activity. However, the stiffness worsened over time, resulting in difficulty in walking and performing daily tasks. Physical examination revealed stiffness over muscles of neck, shoulder and abdominal regions. He had a robot like gait. Most of the neurological examination, including cranial nerves, motor and sensory examination, was unremarkable. Based on the history and physical examination he appeared to be suffering from stiff person syndrome. The pertinent baseline investigations were normal (Table 1).

Table 1: Pertinent baseline investigations

<table>
<thead>
<tr>
<th>Laboratory Investigation</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>CRP</td>
<td>2.35 U/L</td>
</tr>
<tr>
<td>ESR</td>
<td>2 mm 1st hr</td>
</tr>
<tr>
<td>Calcium</td>
<td>9.4 mg/dL</td>
</tr>
<tr>
<td>25 Hydroxy Vitamin D</td>
<td>36.2 ng/mL</td>
</tr>
<tr>
<td>TSH</td>
<td>2.18 micro IU/L</td>
</tr>
<tr>
<td>CPK</td>
<td>199 U/L</td>
</tr>
<tr>
<td>Aldolase</td>
<td>7.0 U/L</td>
</tr>
<tr>
<td>X-Ray Lumbo-sacral spine</td>
<td>Unremarkable</td>
</tr>
<tr>
<td>MRI brain with contrast</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Anti-GAD antibodies were sent which were found to be markedly elevated to a level of > 2000IU/mL. To further confirm our diagnosis, Electromyography (EMG) was done, which showed continuous motor activity in agonist and antagonists muscles, subsiding on the administration of Intravenous diazepam 5 mg (Figure 1).

Figure 1: EMG showing a) continuous motor activity which b) subsided on giving 5 mg IV diazepam.

The patient was treated with Diazepam 2.5mg PO qHS and Baclofen 5mg three times daily. On a two week follow up visit, the patient showed improvement in pain, stiffness and functional status. A Rheumatology consult
Table 2. Dalakas Criteria

<table>
<thead>
<tr>
<th>Dalakas criteria for diagnosis stiff person syndrome</th>
</tr>
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<tbody>
<tr>
<td>Episodic stiffness of the muscles, mostly involving the axial muscles, leading fixed deformity</td>
</tr>
<tr>
<td>Superimposed painful spasms elicited by triggers such as noises, emotional stress and tactile stimuli</td>
</tr>
<tr>
<td>Absence of neurological or cognitive impairment</td>
</tr>
<tr>
<td>Confirmation of continuous motor activity by electro-myographic findings (EMG) (subsides with diazepam*)</td>
</tr>
<tr>
<td>Positive serology for GAD65 or amphiphysin autoantibodies</td>
</tr>
</tbody>
</table>

*not a part of Dalakas criteria, but commonly included in the diagnostic criteria


**Conflict of interest:** Author declares no conflict of interest.

**Funding disclosure:** Nil

**Author’s contribution:**
- **Dr. Zoya Sajjad:** Study concept and design, data collection, data analysis, manuscript writing, manuscript review
- **Dr. Anam Abrar:** Data collection, data analysis, manuscript writing, manuscript review
- **Dr. Zafarullah:** Manuscript writing, manuscript review
- **Dr. Aasim Rehman:** Data analysis, manuscript writing, manuscript review
- **Dr. Wasim Tariq:** Manuscript writing, manuscript review
- **Dr. Arsalan Ahmed:** Data analysis, manuscript writing, manuscript review