Neglect-Like Symptoms in Complex Regional Pain Syndrome: Results of a Self-Administered Survey

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Abstract
Reflex sympathetic dystrophy (RSD), recently reclassified as a complex regional pain syndrome, type I (CRPS-I), is best known for its disabling sensory symptoms, including pain, allodynia, and abnormal skin temperature. Yet, motor dysfunction is common in CRPS and can result in major disability. In addition to weakness of the involved limb, CRPS patients may develop symptoms akin to a neurological neglect-like syndrome, whereby the limb may feel foreign (“cognitive neglect”) and directed mental and visual attention is needed to move the limb (“motor neglect”). Members of the patient support group, the Reflex Sympathetic Dystrophy Syndrome Association (RSDSA), were mailed a questionnaire inserted in their newsletter which inquired about the presence of these neglect-like symptoms; in addition, a separate medical history questionnaire was included to assess adequate documentation for the diagnosis of CRPS. A total of 242 patients returned the questionnaire but only 224 of the questionnaires were analyzed; 15 were excluded due to inadequate documentation of CRPS and 3 were excluded due to non-limb involvement. Eighty-four percent (84%) of these respondents endorsed the presence of at least one neglect symptom and 47% indicated they had both “cognitive” and “motor” neglect symptoms. Of interest, approximately 33% of respondents spontaneously wrote comments regarding the significant disability due to these neglect symptoms and the difficulty explaining these unusual symptoms to their health care providers and family. This patient survey confirms the presence of neglect-like symptoms in a subset of CRPS patients. Neglect-like symptoms need to be addressed and validated by health care providers.

Key Words
Reflex sympathetic dystrophy, complex regional pain syndrome, neglect

Introduction
Reflex sympathetic dystrophy (RSD) has recently been renamed complex regional pain syndrome, type I (CRPS-I) by the International Association for the Study of Pain (IASP). This syndrome is most commonly considered a chronic pain disorder, as pain is the most com-
mon complaint voiced by its sufferers. In addition to localized pain, CRPS patients have, by definition, altered skin temperature, sudomotor activity, and/or skin color within the painful region. Earlier definitions and descriptions of RSD also included motor dysfunction, typically described as a vague weakness, difficulty initiating movement, and tremor, but neither motor symptoms nor signs were included in the diagnostic criteria posited by a consensus panel. However, several recent validation studies of the CRPS criteria have demonstrated the importance of motor symptoms and signs in CRPS, differentiating CRPS from other chronic neuropathic pain syndromes.

Classical teaching has been that motor symptoms and signs in CRPS (RSD) are secondary to voluntary guarding, that is, patients are thought to purposely protect their involved limb in order to minimize pain associated with movement and touching. An alternative hypothesis has been proposed by the authors: some CRPS patients may have an involuntary neurological neglect-like behavior, whereby patients perceive their involved CRPS limb as not part of their being and feeling foreign to them (we will term this “cognitive neglect”) and, in addition, may need to focus mental and visual attention in order to voluntarily move their limb (“motor neglect”).

The aim of this study was to determine the frequency of neglect symptoms among patients with CRPS.

Methods

A two-part questionnaire was sent to members of the Reflex Sympathetic Dystrophy Syndrome Association (RSDSA) along with a regularly mailed quarterly RSDSA Newsletter. (The RSDSA is a nonprofit, nationally based patient support group.) In an attempt to assess adequate documentation of diagnosis, one part of the questionnaire assessed patient demographics, medical history, and overall symptoms, which was reviewed by an experienced neurologist (B.S.G.). Patients were required to have adequate responses that would suggest the diagnosis of RSD in order to be included in analysis; the data were collected prior to reclassifying the disorder as CRPS, but all patients included in the analysis would likely meet current CRPS criteria based on their responses. The data included a history of an inciting traumatic event, involvement of a limb, chronic pain, and a description of dysautonomic symptoms in the area of pain.

The main part of the questionnaire was entitled “Neurobehavioral Questionnaire” (see Appendix 1) and had 5 components assessing the nature and duration of symptoms, the dominant hand, and part of the body affected. A blank space was provided for personal comments.

Four neglect-like symptom statements were developed based on the senior author’s (B.S.G.) personal clinical experience with CRPS patients. The patients were asked to check each statement that was true for their affected limb. Two statements assessed the presence of motor neglect symptoms: “If I don’t focus my attention on my painful limb it would lie still, like dead weight,” and “I need to focus all of my attention on my painful limb to make it move the way I want it to.” Two other statements assessed the presence of cognitive neglect: “My painful limb feels as though it is not part of my body,” and “My painful limb feels dead to me.” A fifth statement assessed for the presence of involuntary movements: “My painful limb sometimes moves involuntarily, without my control.”

Results

Subjects

A total of 242 patients (10%) returned the questionnaires. Fifteen patients’ questionnaires were excluded from analysis due to inadequate documentation for the diagnosis of RSD (CRPS).

The average age of the respondents was 42.0 years (range 19–81 years, SD = 11.6). Fourteen percent were male and 86% female. CRPS involved the upper extremity in 45% of patients and the lower extremity in 55%. The average duration of CRPS symptoms was 53.1 months (range 1–216 months, SD = 41.5).

Questionnaire

One hundred and eighty-eight of the 224 (84%) patients endorsed at least one of the four neglect-like symptom statements. Forty-seven percent (105/224) of the patients endorsed both motor and cognitive neglect state-
ments. With regard to the individual neglect statements: 60% agreed with the cognitive neglect statement “My painful limb feels as though it is not part of my body;” 56% with the motor neglect statement “I need to focus all of my attention on my painful limb to make it move the way I want it to;” 42% with the motor neglect statement “If I don’t focus my attention on my painful limb it would lie still, like dead weight;” 39% with the cognitive neglect statement “My painful limb feels dead to me.” Sixty-eight percent acknowledged the presence of involuntary movements of their affected extremity. No statistical correlation was found between the presence of neglect and right versus left nor dominant versus nondominant side affected.

Approximately one-third of respondents wrote in the “comments” section. They expressed difficulty in discussing these neglect-like symptoms with their health care providers and family members. These patients also wrote of their fearfulness and shame with regard to these unusual neglect symptoms. Many expressed gratitude in validating their symptoms and helping them realize that these neglect symptoms are not unique to them and are not a sign of psychiatric disturbance (e.g., “I am not going crazy.”).

Discussion

The majority of CRPS patients in this survey acknowledged the presence of neglect-like symptoms in their afflicted limb. In fact, a dramatic 84% of 224 patients endorsed at least one symptom of motor or cognitive neglect and 47% endorsed statements reflective of both motor and cognitive neglect. The presence of neglect-like symptoms among individuals with CRPS has only recently been described in one prior published case series by the authors. In that case series, we reported that the CRPS patients perceived their CRPS limb as not part of their being (feeling “foreign” to them) and also needing to focus mental and visual attention in order to voluntarily move their limb. We term the former phenomenon “cognitive neglect” and the latter phenomenon “motor neglect.” The presence of motor neglect can also be assessed on examination, whereby the patient is asked to look away from the involved CRPS limb and move the limb with his or her best effort (lack of visual attention) and then look directly at the limb and perform the same maneuver. If the initiation speed, amplitude, and frequency of the limb’s movements are increased with visual attention, then the presence of motor neglect is confirmed. Our clinical impression has been that both cognitive and motor neglect is present in a subset of CRPS patients. The results of this survey indicate that neglect is an important symptom in a subset of CRPS patients.

If neglect is apparently so common in CRPS, then why has it only recently been described? There are several possible reasons. First, RSD, and now CRPS, are considered first and foremost chronic pain disorders. Patients are more likely to complain of a “positive symptom” like pain than a “negative symptom” such as neglect. Second, as commented by many of the respondents to this survey and by patients we work with, these neglect symptoms are perceived as bizarre, unusual, difficult to describe, and discounted by their health care providers, causing many patients not to discuss their neglect and to question their symptom validity. Some even question their sanity. Third, many pain physicians are trained anesthesiologists and perhaps are not educated about neurological neglect symptoms. Fourth, while motor symptoms have been described in classical descriptions of RSD, it has usually been assumed that lack of movement of the involved limb was due to voluntary guarding of the limb for fear of worsening pain with movement and accidental touching. One prior study did report motor dysfunction in RSD, including the motor neglect sign of “inability or difficulty in initiating movement,” and described a cognitive neglect symptom (“My mind tells my hand to move, but it won’t.”), but ascribed these findings to a “movement disorder” with a possible spinal cord origin.

Interestingly, other data may also suggest the presence of neglect in CRPS. For example, a recent prospective CRPS criteria validation study identified the presence of motor neglect as statistically improving the diagnostic accuracy of CRPS. In addition, functional brain imaging studies (positron emission tomography [PET] and fMRI [functional magnetic resonance imaging] observed decreased activity in the contralateral thalamus in CRPS patients. PET studies of patients with well-defined ne-
glect following brain injury have demonstrated significant focal depressions in the thalamus, as well as the cingulate cortex, prefrontal, and premotor cortex.\textsuperscript{9}

The results of this study need to be confirmed in prospectively examined patients. A limitation of this study is that the subjects are members of a self-referred national support group who claim to have RSD. Even though one of the authors (B.S.G.) reviewed medical data supplied by the patients, diagnosis of CRPS requires direct physical examination, which could not be performed in this study. In addition, selection bias may exist in this study’s patient population, especially considering the extremely low response rate, making generalized interpretation of these data very tenuous.

Nevertheless, this study’s findings regarding the frequent endorsement of neglect-like symptoms in questionnaire responders, in conjunction with our clinical experience, suggest that neglect may be an overlooked but important symptom in some CRPS patients. Neglect can cause both significant physical disability and also can be quite distressing due to its unusual nature. The symptoms and signs of neglect need to be studied prospectively in larger cohorts of CRPS patients. We believe that every CRPS patient should be assessed for the presence of neglect. If present, these patients may significantly benefit from having their neglect symptom validated by the health care provider.

\textbf{Acknowledgment}

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\textbf{References}


\textit{(See Appendix 1 on the following page.)}
Appendix I

Neurobehavioral Questionnaire

I. How long have you had pain in the involved limb?

II. With what hand do you normally write (before pain)? LEFT RIGHT

III. What part of your body is affected with pain? (Circle all that apply)

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<th>OTHER (describe)</th>
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<td>HAND/ARM</td>
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IV. CHECK ALL of the following statements that are TRUE:

1. ____ If I don’t focus my attention on my painful limb it would lie still, like dead weight.
2. ____ My painful limb feels as though it is not part of the rest of my body.
3. ____ I need to focus all of my attention on my painful limb to make it move the way I want it to.
4. ____ My painful limb sometimes moves involuntarily, without my control.
5. ____ My painful limb feels dead to me.

V. If you have further comments with regard to these questions, please comment:

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_____________________________________________________________________
_____________________________________________________________________